Systemic RV in Hypoplastic Left Heart Syndrome After Surgical Palliation

The study by Khoo et al. (1) and the subsequent editorial (2) were read with great enthusiasm and some sense of skepticism. The study comes from 2 well-known pediatric cardiac centers, and the surgical results of the study appear to be commendable. But the authors' conclusion of systemic right ventricle (RV) with an "LV [left ventricular] like contraction" pattern appears to be too much of a generalization and may be far from the target and truth. It is akin to a "pot-bellied" individual trying to do a ballet dance or a computer trying to get emotional intelligence. When we consider the differences between the RV and LV with regard to embryology, anatomy, physiology, and hemodynamics (3–5), it is inconceivable that the RV could adapt to an LV type of contraction. An RV without a middle, circular layer of muscle fibers and without twist or torsion, translation, or radial thickening cannot be compared with the LV.

In the RV, the anatomy of the conus (infundibulum), the "mighty midget" of Van Pragh et al. (6) that is absent in the LV, deserves due consideration while the function of the RV is evaluated by any imaging technique. The unique blood supply pattern (7,8) and the importance of the interventricular septum cannot be underestimated. The extreme difficulty of getting the RV in a 4-chamber view is well appreciated. In the example given, Figure 2, a reasonable image was obtained; it is intriguing that the septum was convex toward the systemic RV. It is interesting to note how far the septum is functionally incorporated into the RV for its systolic function.

A comparison of the pathological data from 3 patients dying within 1 month of the Norwood/Sano palliation with those from the 7 patients dying within the interstage period may throw light on some of the changes in the RV muscle, specially the "circumferential fibers" running parallel to the atrioventricular groove merging with the superficial fibers of the LV (9) and the changes in the conus muscularis. The different patterns of coronary supply may explain the ischemic theory (the post-systolic strain) observed by the researchers. In the mechanism of RV contraction, apart from the inward movement of the free wall ("bellow effect") and the contraction of the longitudinal fibers, the traction of the free wall at the points of attachment to the LV needs to be considered (10). The LV, however small in hypoplastic left heart syndrome, may have a role to play in RV function. Ventricular interdependence cannot be underestimated.

Not withstanding the above considerations, speckle track images to study the RV strain, strain rate, and contraction synchrony will be a great boon to follow up the surgically corrected "little wonders." Powerful imagination is needed to conceive of an RV that can adapt like the LV. At this stage of understanding, we can only have guarded optimism and expectation for the continued performance of the systemic ventricle. The RV adapts better to volume overload than to pressure overload; gradually the RV shows decreased systolic reserve and low cardiac output, which lead to exercise intolerance and fatigue in later life.

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